ASTHENIC BULBAR PALSY.[1]

By Charles W. Burr, M.D.,
NEUROLOGIST TO THE PHILADELPHIA HOSPITAL,

AND

D. J. McCarthy, M.D.,
ASSOCIATE IN CLINICAL MEDICINE IN THE WILLIAM PEPPER LABORATORY OF CLINICAL MEDICINE.

(*From the William Pepper Clinical Laboratory, Phœbe A. Hearst Foundation.*)

We will give no historical account of asthenic bulbar palsy because Dr. Henry Campbell and Mr. Edwin Bramwell have only recently, in the summer number of *Brain*, published a careful summary of reported cases, and have made a thorough critical study of the literature. It will suffice to say that the first case was reported by Wilks in *Guy's Hospital Reports* in 1877. Wilks' patient was a stout, well-looking girl who could scarcely walk. The weakness seemed to be due rather to lethargy than to palsy. She spoke slowly, and had slight nystagmus. A month later many of the symptoms of bulbar palsy appeared, coming on rapidly in about three days. She spoke indistinctly, swallowed with difficulty, and was unable to cough. The limbs were not palsied. Soon breathing became difficult, and in a few hours she died. In the then state of knowledge it would have been proper to have expected to find organic bulbar disease. None was present. Careful macroscopical and microscopical examination revealed nothing abnormal. In 1879 Erb published a similar case, and from that time till 1886 nothing more was written upon the subject. About sixty cases have been reported, with twenty-three deaths and seventeen necropsies. These figures give a false impression of the death-rate, and the high percentage is due to the fact that neurologists are prone to report fatal cases, and many nonfatal cases are probably not diagnosed. At first it was thought the disease affected only muscles innervated from the bulb, but in recent years it has been found to be more wide-spread in bodily distribution, and now it is usually described under the title of myasthenia gravis.

For our own work we propose to relate a case that came to necropsy, to discuss another which, though resembling the first in many ways, yet differs enough to make its proper classification doubtful, and to speculate a little, for the matter is still speculative, as to causation.

We are indebted to Dr. Morris J. Lewis for the early history of the first case.

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[1] Read at the meeting of the College of Physicians of Philadelphia, November 7, 1900.

A woman, aged nineteen years, and married, came to the dispensary of the Infirmary for Nervous Diseases on April 26, 1899. Her family history was negative, her personal history unimportant, except that ever since her marriage, in 1897, she had had much trouble and grief —an unwished-for pregnancy and a brutal husband. In January, 1898, an abortion, probably induced, occurred. She felt no immediate serious ill effects, but about a year later began to suffer from numbness in the right hand, and later in the left hand, legs, and back. She could feel touch, pin-pricks, and the contact of objects without difficulty. The numbness was entirely subjective. Her condition remained the same until a few weeks before she came to the dispensary. She then rapidly grew weak in the arms and legs, so that she could not easily go up or down stairs, and sometimes fell when walking on a level surface.

Examination. She walked a little stiffly and as well with her eyes closed as open. Station was good. The pupils responded to light. The arms and legs were weak. The dynamometer registered fifty in either hand. The knee-jerks were exaggerated, quick, and equal. There was no ankle clonus. The elbow-jerk was exaggerated. She said she could not feel a sharp prick on the hands or legs, and only slightly on the face. She was, however, able to pick up a small piece of paper even when blindfolded. She could distinguish between hot and cold, but said she felt them but slightly. There was a slight tremor of the hands and head. The eyelids trembled and drooped somewhat, covering about half of the pupil. The lids did not come into close apposition on shutting the eyes. She could raise the lids, but not hold them up long. The tongue was tremulous, and when protruded turned toward the left. The respiration rate was twenty-six. The heart was normal. The sphincters were under complete control. There was slight contraction, but no reversal of the color fields. She was easily hypnotized.

She returned to the dispensary a few times, and then was lost sight of until her admission to Dr. Burr's wards at the Philadelphia Hospital on April 23, 1900.

Examination. A young woman of small frame, moderately well nourished, but somewhat pale. The head fell slightly forward upon the chest, and the eyelids drooped, partially covering the pupils. The face lacked expression. She could lift the head, but said it tired her very much to hold it up. She could not close the eyelids tightly, nor open them completely; there was but little power of movement in the frontal muscles, and she could not wrinkle the eyebrows. Her gait resembled that of a person convalescing from some serious acute disease. The movements of the arms were weak. There was no distinct palsy anywhere. Speech was slow and low-voiced—not aphasic, not paralytic, but simply weak. Mastication was slow, and she complained that eating tired her. Her whole manner and bearing showed languor and weariness, but that something more than mere neurasthenia was present was proven by the fact that the amount of weakness varied greatly in different groups of muscles, being most marked in those innervated from the bulb. The knee-jerks were large, but not spastic, and were easily exhausted. Neither ankle nor patella clonus was present. Babinski's reflex was absent; indeed, the plantar reflex was normal. There were slight but quite constant choreiform movements in the face and arms. There was no muscular atrophy of the lips, face, or tongue, and of course none in the extremities. She had difficulty in swallowing, solids seeming to stick in the throat and liquids making her cough. She still complained of numbness in the extremities, but felt touch, pain, and heat and cold well. Dr. Charles A. Oliver examined her eyes and reported: "Vision in each eye is reduced one-half to one-third, that of the right being slightly improved by pin-hole. Pupils are equal in size. Irides respond to light, accommodation and convergence. Extra-ocular movements are very much impeded, especially in outward direction, and more marked to the left. Palsy of convergence in association with a variety of ataxic movements. The eye-grounds are healthy, the fields of vision somewhat concentrically diminished. The patient is unable to close the lids, the action of the left orbicularis being more marked." Her condition remained the same until May 13th, when the difficulty in swallowing suddenly became very great, the temperature rose to 101° F., dyspnœa appeared, the pulse was rapid and weak, cyanosis and coma developed, and she died the next morning.

The necropsy was made the following day; it revealed nothing. The thoracic and abdominal organs were normal. There was a small calcified focus of tuberculosis at the apex of the left lung, and the spleen was chronically enlarged and its capsule much thickened. The kidneys and adrenals were not diseased in any way. The right ovary was cystic. The thymus gland was enlarged, but there was no other evidence of lymphatic diathesis. The uterus contained a three or four-months' old fœtus. The brain, after hardening in formalin, weighed 1250 grammes. Neither it nor the meninges showed any change on gross examination. The cerebrum was of full size, but the pons and medulla were distinctly smaller than normal. The pons was one-third less in diameter than others which had been hardened in the same way. The spinal cord was unusually broad in the lumbar region, and after removal of the dura showed a median dorsal fissure starting at the third lumbar segment and extending to the fifth. At first sight there appeared to be a distinct bifurcation of the cord, but section revealed a persistence of the posterior median fissure extending down to the commissure. The other regions of the cord were normal.

Microscopical Examination. Serial sections of the cord, medulla, pons, and floor of the third ventricle were stained by the Marchi, Nissl, Weigert, carmine, and nuclear stain methods. In the cord no pathological changes were found. In the medulla there were distinct chromotalytic changes with swelling and displacement of the nuclei in the cells of the upper nucleus of the tenth nerve (nucleus terminis vagi). The other bulbar nuclei, including the twelfth and nucleus ambiguous, were perfectly normal. We expected possibly to find changes in the seventh and third nuclei, but they were likewise normal. Sections of the cortex revealed no changes by any of the above-named methods. Sections from all the cranial nerves were examined. By the Marchi method black dots were seen scattered here and there throughout the twelfth, tenth, eighth, and fifth nerves; but inasmuch as similar appearances are seen in healthy nerves, or at least in nerves which have performed their functions well, we attribute no significance to them. Examination of the tenth nerve stained with carmine showed an atrophy of some of the nerve fibres, giving the appearance of a sclerosis. In the tenth and twelfth nerves some swollen axis-cylinders were seen. The muscles showed no pathological changes on microscopical examination.

There are two important questions in the study of this affection: Where is the disease located, and what is its nature? Morbid anatomy gives no help in answering these questions. In the larger number of necropsies no lesion at all has been found, and in the others the changes have been slight and indefinite, similar to those in our case, and of such a nature as not to prove, scarcely to suggest, that they had anything to do with the symptoms. We are compelled, therefore, to fall back upon general physiological and pathological laws, and to draw inferences from analogous affections. The disease must of course be seated in either the muscles or the motor neurons, and if in the latter it must affect either the upper motor neuron from the cerebral cortex to the bulb or cord, or the lower neuron from the bulb and cord to the periphery or some one part, cell body, or axon, of either. The symptoms are so predominately motor that there is no need to look for disease outside the motor apparatus. The disease has been thought to be muscular, but there is much evidence against it. In a large number of cases it has been found that on applying a tetanizing (faradic) current to the muscles at first a brisk contraction is produced, which gradually becomes feeble, and finally ceases, to reappear if the muscle is allowed a period of rest. With the galvanic current, on the contrary, no such exhaustion of muscular contractility occurs. Now faradism causes contraction essentially by acting through the nerves, galvanism by both the nervous system and the muscle directly. Further, Dr. Farquhar Buzzard, at the suggestion of Dr. Campbell, made the following experiment: A moderate galvanic current was applied to the biceps muscle, and a contraction obtained. The muscle was then faradized until it gave no response to a strong stimulus. Then it was tired out by making the patient flex the elbow against resistance, exerted until all power of flexion was lost. On applying the same strength of galvanic current as used at first an excellent contraction was obtained. Finally, on again applying the faradic current, the muscle was found to be still irresponsive. This experiment points strongly against disease of the muscle. Again, we know of no other affection due to disease of the muscles producing analogous symptoms, whereas organic disease of the bulb is not a little similar. Sudden death, a not infrequent thing in asthenic bulbar palsy, is not a symptom of muscular disease. There are several reasons for believing that the upper motor neuron is not affected. The myasthenic reaction, the exhaustibility of the knee-jerk, the pharyngeal palsy, the dyspnœa, the cardiac palpitation, all are evidence against cerebral disease. There is but little direct post-mortem evidence in favor of the bulb as the seat of the disease. The slight changes that have been found were in it and in the cranial nerves. These changes, though almost certainly insufficient to cause the symptoms, may indicate the action of a toxin. The symptoms certainly point toward the bulb as the part first and most seriously affected. Considering everything we may say with comparative safety that the affection is one of the lower motor neurons, but whether the cell bodies or the axons are first and most affected cannot as yet be determined. The primary seat of disease may be in the motor muscular end-plates.

As to the nature of the disease but little is known. It looks like a toxin disease. It frequently follows some mycotic affection. It kills without visible wound. That poisons arising within or without the body may cause death without producing any discoverable lesion goes without saying. That there are diseases without a visible anatomical basis must be admitted, notwithstanding the dogma that there is never perversion of function without alteration of structure. In our case, as in a few others, we are inclined to believe that pregnancy had some causative influence. She was pregnant at the onset, and again when her disease became manifestly serious. Pregnancy may seriously disturb the normal metabolism of the body, as is shown in the kidney of pregnancy, the multiple neuritis occasionally seen, and the greater predisposition of pregnant women to certain diseases. Again, the smallness of the bulb may have had some influence. This was developmental, not secondary, not due to any acquired disease, not caused by the shrinking of an old sclerosis. What nervous elements were present were normal, but they were fewer than usual, they may have been dynamically weaker. It is possible that the ill-developed bulb was unable to withstand the stress of pregnancy.

The second case is made more difficult to understand by the presence of a singular type of anæsthesia. Sensory symptoms of any kind have been but little pronounced in the cases of asthenic bulbar palsy heretofore observed. Occasionally there is some little aching at the back of the neck and in the shoulders, with, it may be, numbness in the arms. True tactile anæsthesia has never been seen. In the case we are about to relate a very interesting form of anæsthesia was present, namely, asteriognosis, the inability to recognize objects by touch, though simple tactile sense is preserved. This symptom makes the proper classification of the case doubtful; but as it more closely resembles asthenic bulbar palsy than any other disease, we place it tentatively there. It would be interesting if in the future there should be met with cases of anæsthesia in the distribution of the sensory cranial nerves causing a condition comparable or at least analogous to the motor disease, bulbar palsy. This is a speculation, of course, but it is possible that such a condition may sometime be discovered. Our case then would be a connecting link. The history is as follows:

A young woman, aged twenty-four years, well educated and of excellent intelligence, with a good family and personal history, came to Dr. Burr, in March of this year, complaining of general neurasthenic symptoms and marked pseudo-emotionalism. She dated her illness from an attack of grippe which occurred in June, 1899. Whether this attack was one of influenza vera caused by the bacillus of Pfeiffer, or the so-called influenza nostras of unknown causation, could not be determined. At all events she never completely recovered. She was tired, languid, weak, and subject to spells of crying unaccompanied by any distressing emotional feeling. She did not regard her condition as serious, accepted no medical treatment, and grew steadily worse. A week before she came under observation her left arm began to be numb, and she had trouble in picking up small objects. A day or two later the numbness extended to the left leg, and still later to the right side. At the same time her neck felt somewhat stiff and there was severe pain in the occiput. She had slight vertigo, blurred vision, and occasional diplopia.

Examination revealed a spare, rather pale young woman. She was very emotional, bursting into tears without any cause. In the midst of her weeping she would say in the most matter-of-fact way that she did not know what she was crying about, and would go on talking about matters and things in general, paying but little attention to the lachrymal flood. Gait and station were normal; the knee-jerks were normal. Anæsthesia was absent. There was no deformity of the spine, nor pain on pressure anywhere in its length. Her condition was not regarded as serious. A week later, however, she suddenly developed palsy of the right face, involving the entire side. This was not an hysterical spasm, but a true palsy. A few hours later there came on excruciating pain in both ears, which lasted throughout the night, and required morphia for its relief. When re-examined there were found a little drooping of the eyelids, a little trouble in swallowing, slightly nasal voice, weakness of the muscles of mastication on both sides, but the right facial palsy had improved. On rising she would walk well for a few minutes, but soon would stagger and be compelled to sit down. All movements of the left arm were present, but all were weak, and soon she could not move the fingers or wrist against the slightest resistance. She could not squeeze the dynamometer at all. Power in the right arm and hand was a little greater. At no time was there complete loss of power in the arms. She could always make unresisted movements, but could not even lift a spoon, could not feed herself, and even unresisted movements soon tired her so much that she could not continue them. She could not write a word, could not grasp a pen strongly enough to hold it. In bed the movements of the legs were done weakly, but even at the worst she could stand and walk a little. The legs were never so seriously affected as the arms. She could move the tongue in all directions, but complained that the tip felt as if it had been burnt. There was no muscular wasting anywhere. The electrical reactions were normal, but unfortunately no examination was made for faradic exhaustibility. She had frequent attacks of pain in the occiput and back of the left ear, especially if she sat up in bed with the head unsupported. Palpation of the neck and head did not increase the pain at the time, but after an examination pain was apt to occur. No deformity of any kind was discoverable in the occipital region or in the cervical spine. There was no spasm of the muscles of the neck.

The knee-jerks were normal, and ankle clonus was not present. Touch, pain, and temperature-sense were normal on the arms, legs, and face; but later, when the power in the arms had returned and she could grasp well, it was discovered that she could not recognize objects by touch. This test, of course, could not be made while she was unable to handle objects. Smell and taste were normal throughout the course of the disease.

Dr. John T. Carpenter examined her eyes and reported as follows: "Central vision normal; field for form and color strictly normal. No reversal of the fields. No pathological changes in the eye-grounds. Refraction error; moderate hypermetropia. No diplopia except when red glass interposed, when vertical diplopia results and is constant. Right hyperphoria 2°, which is concomitant, not paretic. Ocular movements normal, though both adduction and abduction are below par (15°—5°). The only important factor elicited was the discovery of constant right hyperphoria." The sphincters were under complete control.

On first seeing the patient I did not regard her as seriously ill, but when the facial palsy appeared, followed by the difficulty in swallowing, the nasal speech, and the great muscular weakness, I began to fear a very serious issue, and believed that there was organic disease in the bulb and upper cervical cord or in the neighboring bones. Events proved this was an error, for after a few months she improved rapidly, and now is quite well.

There is no specific in the treatment of asthenic bulbar palsy. Rest is the most important element. The patient should be put to bed; faradism does harm; galvanism is of doubtful benefit; massage is useful. As to medicines, arsenic, in our experience, is the one drug which seems to be of benefit.

A STUDY OF A CASE OF GONORRHŒAL ULCERATIVE ENDOCARDITIS, WITH CULTIVATION OF THE GONOCOCCUS.

By August Jerome Lartigau, M.D.,
OF NEW YORK CITY,
TUTOR IN PATHOLOGY, COLLEGE OF PHYSICIANS AND SURGEONS, COLUMBIA UNIVERSITY; PATHOLOGIST TO THE SLOANE MATERNITY HOSPITAL; ASSISTANT PATHOLOGIST TO THE ROOSEVELT HOSPITAL.

(*From the Department of Pathology, College of Physicians and Surgeons, Columbia University.*)

Notwithstanding the wide influence of the original assertion of Bumm that gonorrhœal processes remain limited to mucous membranes, the fact has been clearly established that the gonococcus may, under certain conditions, incite the production of a series of lesions in the body tissues generally, as dissimilar in pathological significance as they are varied in their clinical aspects. Such a relationship has been